Annals of Cardiology and Cardiovascular Medicine

Case Report
Published: 19 Apr, 2019

Ebstein's Anomaly with Severe Tricuspid Valve Stenosis Presenting as Acute Embolic Stroke Secondary to Deep Vein Thrombosis

Veena Nanjappa, Hema Raveesh, Sadanand KS and Manjunath CN*

Department of Cardiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research, India

Abstract

Ebstein's malformation is an uncommon congenital heart disease with great variability in the morphology of the tricuspid valve. Its prevalence is 0.3% to 0.5% and less than 10% cases are associated with severe tricuspid valve stenosis. Patients with Ebstein's anomaly have 10% incidence of pre-excitation. We hereby report a similar case of Ebstein's anomaly with asymptomatic cardiac status presenting for the first time at adult age with embolic stroke as a result of paradoxical embolism via ostium secundum atrial septal defect secondary to left leg deep vein thrombosis with pre-excitation syndrome on ECG.

Keywords: Ebstein's anomaly; Atrial septal defect; Tricuspid valve stenosis; DVT

Introduction

Ebstein's malformation is an uncommon congenital heart disease with great variability in the morphology of the tricuspid valve. In the majority of cases, dysplasia and displacement of the tricuspid valve leaflets result in varying degrees of tricuspid regurgitation. In some, however, there may be linear attachments of the antero-superior and mural leaflets of tricuspid valve to the junction between the inlet and trabecular components of the right ventricle, resulting in imperforate type of tricuspid valve or embryologically may arrest later on in development resulting in severe tricuspid stenosis [1].

We hereby report a case of Ebstein's anomaly with severe tricuspid valve stenosis with NYHA functional class II status presenting for the first time at adult age with left hemiparesis as a result of paradoxical embolism via ostium secundum Atrial Septal Defect (ASD) secondary to left leg Deep Vein Thrombosis (DVT) with pre-excitation syndrome on ECG.

Case Presentation

A 32 year old young male presented to the neurologist with transient left hemiparesis and dysarthria and was diagnosed with stroke. He also gave history of painful swelling of both lower limbs of two weeks duration. MRI brain revealed acute infarcts in right basal ganglia and corona radiate (Figure 1); subacute infarcts in right posterior temporal lobe and also chronic infarcts in bilateral periventricular and centrum semi ovale regions. MR angiogram revealed distal middle cerebral artery embolus.

On clinical examination he had bilateral tender swelling of both lower limbs (left > right) and vitals were stable with no perceptible murmur on cardiac examination. He had central cyanosis with saturation of 88% at rest and prior to his CVA he was in functional class II and was working as a farmer. There was no differential cyanosis. He had pan digital grade II clubbing. On further questioning, he gave no history of cyanosis, palpitations or dyspnea in the past. He was born of a non-consanguineous marriage and had uneventful birth history. There was no history of any congenital heart disease in his siblings. His ECG revealed pre-excitation syndrome with short PR and delta wave suggestive of a possible right postero-septal pathway; his ECHO work up revealed Ebstein's anomaly with 4.5 mm apical displacement of hinge point of tricuspid valve; he also had severe tricuspid valve stenosis with dilated right atrium with trickle of flow seen across the tricuspid valve. Linear attachments of the anterior and the mural leaflets resulted in almost complete separation of the inlet portion from the apical trabecular and outlet portions of the right ventricle. He had a large bidirectional ostium secundum ASD 4.2 cm in size (Figure 2). He had LV global

OPEN ACCESS

Correspondence:
Hema Raveesh, Department of Cardiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research, Mysore, India, Tel: 919886512210; E-mail: hemaraveesh@yahoo.com

Received Date: *14 Mar 2019*
Accepted Date: *12 Apr 2019*
Published Date: *19 Apr 2019*

Citation:
Nanjappa V, Raveesh H, Sadanand KS, Manjunath CN. Ebstein's Anomaly with Severe Tricuspid Valve Stenosis Presenting as Acute Embolic Stroke Secondary to Deep Vein Thrombosis. Ann Cardiol Cardiovasc Med. 2019; 3(1): 1020.

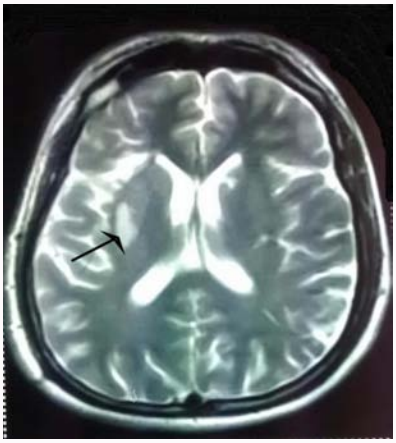

Figure 1: MRI brain: Image of acute infarcts in right basal ganglia and corona radiata; subacute infarcts were seen in right posterior temporal lobe and chronic infarcts were seen in bilateral periventricular and centrum semi ovale regions. MR angiogram revealed distal MCA embolus.

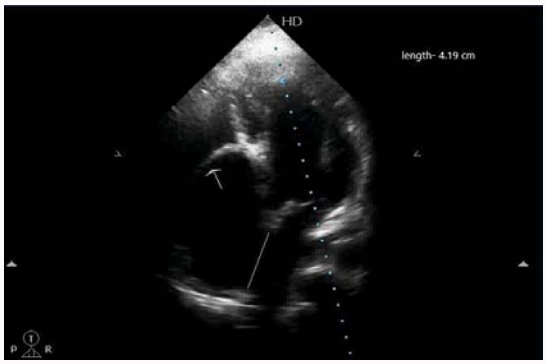

Figure 2: Transthoracic 2D images: Ebstein's anomaly in four chamber view; apical displacement of tricuspid valve seen, with large ostium secundum ASD.

hypokinesia with EF 40%. There was no Doppler gradient across the right ventricular outflow tract and the pulmonary valve. Right ventricle appeared morphologically normal, although small. There was no demonstrable VSD or PDA on transthoracic echo. Since he gave no history of palpitations or syncope and no family history of heart disease he was not considered for electrophysiological study.

His cardiac MRI revealed severe tricuspid valve stenosis with atrialization of right ventricle suggestive of Ebstein's anomaly (Figure 3). Annular diameter of tricuspid valve was 3 mm (Normal is: 2.8 cm to 3.1 cm) suggesting severe tricuspid valve stenosis. His RV was very small.

He also had contrast induced nephropathy following MR angiogram. Venous Doppler of lower limb revealed tibial vein DVT extending till great saphenous vein. His thrombophilia work up and Antinuclear Antibodies (ANA) were negative. His hemoglobin was 17% with hematocrit of 59%. He was started on parenteral and overlap-oral anticoagulation till therapeutic INR was achieved. Cardiac catheterisation study was not considered in view of his extensive DVT. His Celermajer score was not conducive for intra cardiac repair.

Discussion

Ebstein's anomaly is an uncommon congenital heart defect with a

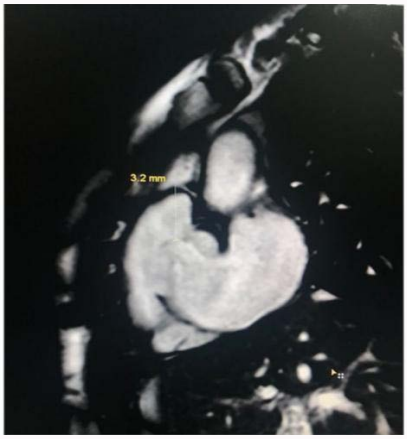

Figure 3: Cardiac MRI shows severe tricuspid valve stenosis-Annular diameter of tricuspid valve 3.2 mm. Atrialization of right ventricle seen.

prevalence of 0.3% to 0.5% [2], occurring in 1 per 200,000 live births. The following anomalies have been reported that may be associated with Ebstein's anomaly: Atrial septal defect (90%), anatomic or functional pulmonary atresia (30%) and ventricular septal defect (less common). Its association with an imperforate tricuspid valve and tricuspid valve stenosis is seen in only less than 10% of cases [3,4]. This imperforation or stenosis occurs due to the excessive redundancy of the anterior leaflet, with fusion of the commissures. Partial fusion of developing tricuspid valve leaflet components results in tricuspid stenosis and complete fusion results in tricuspid valve atresia [1,5].

Tethering and displacement of the tricuspid leaflets result in a very peculiar echocardiographic appearance, with an "empty" tricuspid annulus and membranous separation of the inflow and the outflow portions of the right ventricle. This anatomic condition has been and continues to be the subject of discussion, as functionally it may be classified as a subtype of tricuspid atresia, but embryologically it belongs to the spectrum of Ebstein's anomaly [6]. The first cases were described in anatomic specimens by Van Praagh et al. [7] in 1971.

Patients with Ebstein's anomaly have 10% incidence of pre-excitation. Our patient had asymptomatic pre-excitation syndrome with demonstrable delta wave in resting ECG. He was however not subjected to stress test in view of his acuity of illness.

Celermajer et al. [6] reviewed 220 cases of Ebstein's anomaly with 1 to 34 years of follow-up. Actuarial survival for all live-born patients was 67% at 1 year and 59% at 10 years. Predictors of death were echocardiographic grade of severity at presentation (relative risk increased by 2.7 for each increase in grade), fetal presentation, and right ventricular outflow tract obstruction.

Observation alone is advised for asymptomatic patients with no right-to-left shunting and only mild cardiomegaly. Children who have survived infancy generally do well for several years, and surgery can be postponed until symptoms appear, cyanosis becomes evident, or paradoxical emboli occur. Deliberations about an operation should begin if evidence of deterioration exists, such as progressive increase in right heart size, reduction in systolic function, or appearance of ventricular or atrial tachyarrhythmias. Once symptoms progress to NYHA functional class III or IV, medical management has little to offer, surgical risks increase, and operation is clearly indicated. A biventricular reconstruction is feasible for most patients in an

experienced cardiac centre. However, because of the wide spectrum of anatomic variations in the tricuspid valve, the surgical approach to patients with Ebstein's anomaly needs to be individualized according to the specific morphology found at operation. A one and half ventricle repair can be applied to the failing right ventricle. Heart transplantation is reserved for patients with severe biventricular dysfunction. There are no surgical series data of Ebstein's in India. In the society of thoracic surgeons' congenital heart surgery database data [8], in hospital operative mortality was found to be 3.3% in adults and late mortality of 10%.

Bidirectional Glenn with closure of atrial septal defect was contemplated for this patient in concordance with surgeon at later date.

Conclusion

This case highlights the importance of cardiac evaluation especially in stroke. Congenital heart diseases as a causal pathology need to be kept in mind in the evaluation of stroke in young.

References

1. Wilson AD, Rao PS. Embryology. In: Kambam J, editor. Cardiac Anesthesia for Infants and Children. St Louis: MoMosby; 1994. p. 3-9.
2. Anderson RH, Shinebourne EA, Macartney FJ, Tynan M. Ebstein's malformation and related lesions of the tricuspid valve. Pediatr Cardio. 1987:721-36.
3. Maron BJ, Towbin JA, Thiene G, Antzelevitch C, Corrado D, Arnett D, et al. Contemporary definitions and classification of the cardiomyopathies: An American heart association scientific statement from the council on clinical cardiology, heart failure and transplantation committee; quality of care and outcomes research and functional genomics and translational biology interdisciplinary working groups; and council on epidemiology and prevention. Circulation. 2006;113(14):1807-16.
4. Attenhofer Jost CH, Connolly HM, O'Leary PW, Warnes CA, Tajik AJ, Seward JB. Left heart lesions in patients with Ebstein's anomaly. Mayo Clin Proc. 2005;80(3):361-8.
5. Rao PS. Tricuspid atresia. In: Long WA, editor. Fetal and Neonatal Cardiology. Xth ed. Philadelphia: WB Saunders: 1990.p. 525-40.
6. Celermajer DS, Bull C, Till JA, Cullen S, Vassillikos VP, Sullivan ID, et al. Ebstein's anomaly: Presentation and outcome from fetus to adult. J Am Coll Cardiol. 1994;23(1):170-6.
7. Van Praagh R, Ongley PA, Swan HJC. Anatomic types of single or common ventricle in man. Morphologic and geometric aspects of sixty necropsied cases. Am J Cardiol. 1964;13:367-86.
8. Davies RR, Pasquali SK, Jacobs ML, Jacobs JJ, Wallace AS, Pizarro C. Current spectrum of surgical procedures performed for Ebstein's malformation: An analysis of the society of thoracic surgeons' congenital heart surgery database. Ann Thorac Surg. 2013;96(5):1703-9.